The Nature & Affinities of Chorea.

The evidence that Chorea is a disease of neurotic origin is I think well founded and convincing. In this thesis I propose to adduce evidence to show that it is a mild psychosis or a disturbance in the emotional substratum with motor accompaniments or resultants having close affinities to epilepsy and hysteria.

The common element to all the so-called neuroses is an undue irritability of nerve cells whereby on excitation there is a disengagement of nerve energy in excess of economical requirements.

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This, I premise, as the physiological or possibly the pathological definition of a neurosis. The causative element is therefore considered to be identical, while the symptomatic element is dependent on the area first involved and on the area of the distributed discharge. This definition aids us considerably in forming a conception of the hereditary relationship, transmutability, or interchangeability of the various neuroses. Convulsions in childhood, pavor nocturnus or chorea at 7 years of age, megrim, asthma or epilepsy,

at 14, hysteria a adolescent insanity at 18, may be viewed as but different phases in the neurotic cycle. The disappearance of asthma where hysteria or epilepsy supervenes, the substitution of mania transitoria for an epileptic convulsion, or the fusion of chorea with hysteria prove those affections to be relative quantities and their individuality to be sometimes masked or lost. This transmutation is conspicuous from the hereditary point of view. Epilepsy does not necessarily beget epilepsy: it may be genetically responsible

for asthma, megrim, chorea, hysteria or insanity.

The comparative study of the Symptoms or groups of Symptoms of Chorea and epilepsy establishes an apparent kinship. Acute wild delirium, varying degrees of dementia or at least of mental enfeeblement, transient hemiplegia or hemianaesthesia, speech difficulties or aphasia are alike common to both diseases. Chorea major, a true convulsive explosion, is an approximation to the Status Epilepticus; and I have frequently noticed in Asylum practice

that purposeless or choreiform movements are frequent after an epileptic fit. Epilepsy has been known to supervene on Chorea. Hysteria simulates or even passes into genuine epilepsy. There is a reflex epilepsy and a reflex chorea: the former may be caused by foreign bodies in the ear, nasal polypi &c; the latter by intestinal irritation "Chorea is sometimes eventually lost sight of in hysteria pure and simple'" Add to those clinical facts the contagious element in those affections

(1 Stuges: Chorea p. 64)

and their transmutability and interchangeability by heredity, and the evidence that their causes are similar in nature may be reasonably entertained.

I have collected notes of 20 cases of chorea that I have seen in general practice during the last two years. The families are all personally known to me. Eighteen were females between the ages of 7 and 15 years, the other two were males (one a boy of 8, the other a man of 40.) They were all without exception highly nervous and excitable.

so that I would be justified in inferring that chorea is remarkably partial in its choice of subjects. There were 7 cases of dipsomania in the male parentage. The associated diseases in the direct and collateral lines were infantile convulsions, convergent strabismus, chlorosis, epilepsy (in two), megrim and hysteria. Rheumatism was known in four of the families, but rheumatic fever only attacked two of my choreic patients.

The condition of the palate deserves careful attention as

Shape of palate in man aet. 40 (chorea)

neurotic palate (chorea)

neurotic palate of a woman, aet. 25 (chorea)

neurotic palate (Chorea)

slightly deformed palate in a child (chorea)

Shapes of 5 Palates from cases of Chorea

I think it helps us to identify Chorea as a neurosis, or at least to strengthen the probability of it having such an origin. Clouston having pointed out the frequency of this deformity or abnormality in idiots, in adolescent insanity, and in some other developmental diseases, it occurred to me that Chorea, assuming it to have the same nosological status, ought to or might be expected to show this deformity of the hard palate. I therefore examined all the palates of my patients and took their shape very carefully

and found that 12 out of the 20 had a well marked 'neurotic' palate and that 6 others were slightly 'deformed'. To draw a universal inference from my small number of cases would be rash and illogical; but at the same time it is instructive that so far as I have had opportunities of pursuing the inquiry the percentage of abnormal palates is as high as Clouston has found it to be in congenital and in adolescent insanities. Regarding the morphological significance of this sign Clouston says:- "We

" We must refer the high palate to a bad initial neurotic heredity... The vaulted palate and altered dental arch must be taken with other changes in the head, and especially in the face expression, as one of the morphological indications that show a tendency in the person to whom it belongs and in his family towards developmental neurotic diseases, notably idiocy, congenital imbecility, deformity, epilepsy, adolescent insanity and that organic lawlessness and lack of mental inhibition & weakness of mind that

distinguish the criminal classes.
It thereby is one of the marks of
a family that is tending towards
mental death and extinction.
Taking all the facts into account
it seems proved that the condition
of the palate may be a most
important index of brain development
and of liability to the
developmental neuroses" [1]

We may therefore with reason
accept this as an additional
argument for chorea being
relegated to the group of neuroses.

Among the predisposing causes
of chorea I have been led
by an intimate acquaintance

(1 Clouston: Neuroses of Development. p. 51)

with my own patients to the conclusion that a nervous temperament is a sine qua non. If not overt in the subject it is easily demonstrable in a parent or other near relative. What is the fundamental and physiological significance of a nervous temperament? Is not its dynamical equivalent the definition already given that it is an abnormal excitability of nerve cells in virtue of which a stimulus occasions a disengagement of nerve energy in excess of economical requirements? Does this not help us to understand why in such

subjects a fright or other emotional disturbance should upset muscular control? To substantiate this it may be advisable to review the physiological correlatives of emotion as affecting the muscular system. The word emotion itself, as connoting movement, implies that muscular contraction is its correlative or dynamical equivalent, and shows the profound and ineradicable influence emotion has on the organic life. A faint wave of pleasurable feeling reveals itself by the wrinkling

of the skin at the outer angle of the eye by the orbicularis palpebrarum and the retraction of the angles of the mouth by the risorius: as the feeling rises in intensity the muscular response becomes more diffuse pari passu, involving the other facial muscles, the laryngeal, the forced inspiratory & expiratory, until perhaps the whole system is convulsed in laughter. The sportiveness and dancings of joy, the inability to sit still after the receipt of welcomed news, the weeping

and wailing of grief, tearing the hair from fury agony or despair, the dilated nostrils, the frowning, the set teeth, the clenched fists or the stampings of anger or of pain, the screaming of fright, the trembling of fear or of rage are but a few illustrations of the organic connection between the emotions & the muscles.

"To all appearances a violent emotion may act sometimes in the same way as a strong physical shock to the nervous system, for it may

produce in some instances convulsions, fainting, loss of sensation paralysis & deafness"[1]

The general paralytic, a person of great emotional excitability, is in perpetual motion day and night. The influence of emotion on the vascular system is no less marked. Expectancy or suspense increases the frequency of cardiac pulsation, whilst grief has an opposite effect. Vaso-motor paralysis, fainting, a fatal syncope may also be the consequences

(1 Maudsley: Physiology of Mind P. 360)

of emotional disturbance.

It seems clear therefore that an excitation in the emotional substratum must, as an organic necessity, find expression in muscular movement.

Prior to the onset of Chorea there is usually an apparent depreciation of the general health. All my cases showed some degree of anaemia — ten of them markedly so. This anaemia probably plays an important rôle in the induction of Chorea. It is a well known fact that

anaemia causes many minor mental symptoms E.g. apathy, inaptitude for work, irritability, depression &c. Indeed an anaemic person has a decided proclivity towards emotional disturbance. If anaemia becomes extreme it causes convulsions – as in haemorrhage If loss of blood can produce general convulsions is it wonderful that a certain dgree of anaemia or nutritive deficiency of the blood should produce or tend to produce motor disturbance especially if it should act along with

fear or fright or other emotion which we have seen to have such essential influence on muscular movement?

Another predisposing cause perhaps of some importance (not to mention age, sex or rheumatism) is the great relative weight of the brain to the body in children. The brain has attained its full growth up to a few ounces at the 7th year. We may reasonably assume then that its dynamical equivalent is relatively greater than in the adult. This is

supported by the instinctive fidgetiness and restlessness of children. A child from a nervo-muscular point of view is a very unstable quantity only yielding to the tyranny of its organisation in perpetual romping.

The confinement and strict regime of schools tend to inhibit this physiological outlet of nerve energy and by so doing probably act as a predisposing cause of Chorea

"It is to be remarked that children away from civilization and who are suffered to grow up in their own way

with little or no training are indisposed to take Chorea"[1]

Further, forcing a young brain into a too highly evolved activity by relatively difficult- or advanced lessons must have a deleterious effect on the development- of the highest centres and an evil reflex effect on the emotions.

There is no doubt that fear or fright is one of the most efficient causes of Chorea. It was the alleged cause in 12 of my 20 cases. I was careful not to suggest such

(1 Sturges: Chorea p. 31)

a thing to the parents so that it was spontaneously given.

Its modus operandi is perhaps not difficult to understand. Couched in physiological terms a fright is an explosive lesion in a special area of the emotional substratum.

"Nervous stimulations and discharges consist of waves of molecular change that chase one another rapidly through nerve fibres. Each set of waves while itself caused by the decomposition of unstable nerve matter is a means of decomposing other.

unstable nerve matter, so generating further and often stranger sets of waves which similarly chase one-another into many and distant parts of the nervous System"[1]

This appears to me to be a concise and philosophical description of the proximate pathology of Chorea. The initial explosion (as by fright) is the means of decomposing other unstable nerve centres in the emotional substratum, the physical correlative of this cyclical molecular movement being

(1 Spencers Principles of Psychology Vol I p. 95

that now one set and now another set of muscles is affected, the muscular response varying quantitatively as the strength of wave. Nerve current always travels along the line of least resistance; consequently we find those muscles most involved where there is least inertia to be overcome. In harmony with this the facial muscles in adults are always conspicuously affected, indeed sometimes the only ones that are so. This is a clinical

fact of great significance in studying the etiology of choreiform movement. The fact that the muscles of expression — the muscles of emotion par excellence — are always involved in the adult- is a prima facie argument in favour of the cortical lesion being in the emotional substratum. On the other hand the fact that in young children those muscles often escape is not incompatible with the hypothesis, because at an early age the muscles.

of expression are not in co-ordinated correspondence with the emotions. Further, if an affected group of muscles be held at rest the movement will be transferred to some other muscle or group of muscles. This, if duly considered, seems to show, not that the lesion is in the nerve centre presiding over the particular muscles in question but in a still higher centre, the discharge following the line of least resistance. As further corroborative evidence of the theory that the primary

disturbance is not in the muscle centres but in the emotional. is the fact that the leg muscles are never affected alone. If embolism were the determining cause of chorea would it not be reasonable to expect that the motor area for the leg would sometimes be the only part disturbed by the emboles? Why should an embolus have such a constant and unfailing predilection for the facial muscles in adults, and have no such selective affinity in young children? Why are

idiots so notoriously immune from Chorea? Do they enjoy immunity from Embolism? Is it not more satisfactorily explained by the emotional theory that in their less evolved mentation, in their abortive emotional development, the causative substratum in idiots is awanting. It is true that idiots are prone to sudden outbreaks of passion, but these again are relieved by convulsive explosions. Is Embolism not responsible more for paralysis than for

the contraction of muscles?

Why do coma, hemiplegia or aphasia not occur with greater frequency in choreic children?

As a second attack of chorea always resembles the first in its muscular distribution how happens it that the emboli always block the same arteries at the same site?

Further, it might reasonably be asked if recovery could be so complete after such a gross lesion as embolic plugging?

The fact that fright makes

a lasting impression on a nervous child is irrefragable, so that it may be a considerable time before the nerve cells regain their equilibrium. The mere re-presentation of the fright in the child's mind may serve to make the disturbance persist. Though fright appears to be a very frequent and efficient cause of chorea there are undoubtedly other causative agents alike in kind but varying in degree. Grief, anxiety, worry, depression, &c if long continued

and acting in conjunction with anaemia and the nervous temperament, may each produce that irritability or explosiveness in nerve cells which presumably is the causa causans of Chorea. One of my patients a married woman, aet. 25°, in the early months of a first pregnancy & suffering from a first attack of Chorea, imputed the cause to the depressing and worrying influence of the coal-strike as it threw her husband out of employment

and made their living precarious.

I find that an emotional disturbance of varying degree to be the invariable precursor of the choreic movements. I have always satisfied myself on this point. The child's disposition is changed. She has become fretful, capricious, peevish or unusually timid; sometimes she is irritable; she cries on slight provocation; she is fond of being petted (perhaps the analogue of the longing after sympathy by hysterical females)

she does not take the same lively interest in childish amusements; she is more taciturn than she is wont to be; her power of attention is often impaired. One of my patients a girl of 9 and a smart girl at school had quite forgotten the simplest elements in her multiplication tables. Do not these symptoms justify one in diagnosing a mild affective insanity?

To explicate the final cause of this emotional disturbance is

well-nigh impossible in the present imperfect state of cerebral pathology. The organic conditions necessary for the healthy action of a nerve cell, the quantity and quality of nutrition, the cause and chemical products of the disintegration of nerve matter, the cause & nature of molecular motion are so imperfectly known that, in the want of knowledge of physiological processes it is impossible to fathom the causes of pathological processes. We must therefore be contented with proximate causes.

It appears to me that the necessary conditions for the development of chorea are a certain degree of nervous temperament, the supervention of anaemia and consequent on this, perhaps from deficient nutrition, emotional irritability produced of the motor symptoms.

It is a universally recognised fact that chorea may be contracted by contagion. This surely militates against an embolic or thrombotic genesis! We can find the rationale of its action if we but reflect on the imitative tendency

of children; indeed of the whole human species. "When we fix the countenance in the expression or the body in the attitude which any passion naturally occasions it is certain we acquire in some degree that passion, and if we try while the features are fixed in the pattern of one emotion to call up in the mind a quite different one we will find it impossible to do so".[1]

Is not this its psychological explanation? The mere imitation of facial expression or attitude

[1] Maudsley: Physiology of Mind p. 387.

occasions an emotional response which in a predisposed subject may become deflected in choreiform movement.

Why should Rheumatism produce Chorea? An attack of rheumatism is attended with great and acute suffering. The natural expression of pain is muscular movement — groaning biting the lip, setting the teeth, stamping the feet &c. When several joints are affected the patient's natural motor outlets for the relief of pain are very much curtailed. He feels there is a great tendency for the affected limbs to be

moved by reflex action alone - the points of irritation being in the joints. He also knows that the slightest movements cause extreme suffering, consequently he must exercise inhibitory powers to prevent reflex movement. This must cause anxiety. Fear, lest the limb should be moved and great arthritic pain co-operate to give the initial emotional disturbance which, according to our hypothesis, may be causative of chorea in a predisposed subject.

The tendency to rheumatism is inherited: if there is a choreic tendency also what is

more natural to expect than that chorea should assert itself at a critical time when the emotional equipoise is upset. This modus operandi is conceivable and credible. Why rheumatism should complicate or follow chorea is a much more difficult question to answer than the former, inasmuch, the etiology of rheumatism is not definitely known. In only 2 of my 20 cases did rheumatism supervene on chorea (both in the 3rd week) and, mirabile dictu, those were the only cases that had previously been confined indoors so that

there was no exposure to account for the onset of rheumatism. As far as my experience has gone I have noticed that rheumatism is a much more frequent complication of chorea in hospitals than in private practice. The question immediately suggests itself whether close confinement to the house or to bed has any thing to do with the appearance of rheumatism. My other 16 cases had regular out-door exercise & there was no such complication in any of them. If we assume that Rheumatic fever is caused by some organic element in the blood it is possible to construct a

rational hypothesis. If rheumatism, as is alleged by many, is produced by disturbances in the nutritive and eliminatory functions with the consequent development and retention of Lactic acid or other similar organic irritant in the system, have we not those conditions fulfilled in Chorea? There are digestive disturbances, the emunctories act sluggishly, the perpetual muscular movement impelles the production of a considerable detritus containing a large percentage of Lactic acid; Similar products result from the disintegrating processes going on in the nerve cells. As the eliminating organs are working perfunctorily there probably is a retention in

the system of some metabolic product which we have assumed to be the efficient cause of rheumatism. This would explain the greater tendency for rheumatism to show itself in those cases which are closely confined, as want of open air exercise would retard still more the eliminatory functions. Whether there must be a rheumatic predisposition to determine the result I cannot say, but in both the cases of Chorea under my care complicated with Rheumatism there was such a family taint. On the other hand it is held by some that rheumatism has its origin in the nervous system. As remarked by Clouston the neurotic affinities of rheumatism have yet to be worked out.

Until that is accomplished it will be impossible to reconcile the frequent concurrency or inter-dependency of chorea and rheumatism from a neurotic point of view.

The prophylactic treatment of chorea merits attention. A family physician thoroughly cognisant of the proclivities of his patients and their relatives can often detect in a child's disposition the conditions tending towards the development of chorea. He would thereby recommend much out-door exercise and amusement, short school hours and no hard cramming lessons. In short the indication is to prevent mental overwork

to subdue precocity, to encourage childish sport and every other healthy outlet for that abundant energy so conspicuous in children. Those iniquitous and ubiquitous Ghost stories should be rigidly interdicted. Neurotic children are bad companions for one another.

As regards therapeutic agents iron is indicated in the vast majority of cases. It has more influence over chorea than any other separate drug that I have tried. The general health soon improves after its exhibition, and it appears to expedite recovery.

In a chronic case of over a years duration the regular administration of Bromide of Potassium for 2 months seemed to ameliorate and finally to remove all the symptoms. This drug is perhaps well worth a trial in the treatment of Chorea.
